CASE REPORTS

SPONTANEOUS SPLENIC RUPTURE IN VASCULAR EHLERS-DANLOS SYNDROME: A CASE REPORT MANAGED BY COIL EMBOLIZATION OF THE SPLENIC ARTERY

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Abstract

Vascular Ehlers–Danlos Syndrome (vEDS) is a rare genetic disorder characterized by arterial fragility and a high risk of spontaneous vascular rupture. Splenic artery rupture is an uncommon but life-threatening manifestation of this condition.

We report the case of a 34-year-old man with vEDS who presented with sudden abdominal pain and rapidly progressed to hemorrhagic shock. Imaging confirmed active bleeding from the distal splenic artery. After multidisciplinary evaluation, endovascular embolization via a left humeral approach was performed using multiple coils. Hemostasis was successfully achieved, and the patient showed immediate clinical improvement. He was discharged after 27 days following immunization against encapsulated organisms due to presumed hyposplenism.

This case highlights the crucial role of endovascular techniques in managing vascular emergencies in vEDS, offering a minimally invasive and effective alternative to open surgery. Splenic artery embolization may be life-saving, but long-term surveillance and preventive strategies remain essential, given the risk of future arterial events.

Keywords: *ascular Ehlers-Danlos Syndrome, spontaneous splenic rupture, endovascular embolization, arterial fragility, hemorrhagic shock.*

INTRODUCTION

Ehlers–Danlos Syndrome (EDS) comprises a heterogeneous group of rare inherited connective-tissue disorders associated with abnormalities in skin, joints, and vasculature. Vascular Ehlers–Danlos Syndrome (vEDS), previously termed EDS type IV, is the rarest and most life-threatening subtype because of its strong association with spontaneous arterial dissections and ruptures, particularly in medium-sized arteries. It results from autosomal dominant pathogenic variants in COL3A1 or, less frequently, COL1A1, leading to defective type III or type I collagen synthesis, structure, or secretion.[1-4]

Approximately 80% of patients experience a major vascular complication by age 40[5].Splenic artery pathology is reported in 7.9% to 21% of patients with vEDS, compared with 0.8% in the general population. Managing arterial complications in vEDS remains challenging because of extreme tissue fragility and the high risk of catastrophic intraoperative bleeding.[3-5]

We present a case of spontaneous splenic artery rupture in a patient with vEDS, illustrating the difficulties in therapeutic decision-making and the role of endovascular embolization as a safe and effective option.

CASE REPORT

A 34-year-old man with a known diagnosis of vEDS presented to the Emergency Department with sudden, intense abdominal pain, without preceding trauma. Although he was hemodynamically stable on arrival, he rapidly deteriorated into hemorrhagic shock within two hours.

Immediate resuscitation with intravenous fluids, massive transfusion, and peripheral vasopressors was initiated.

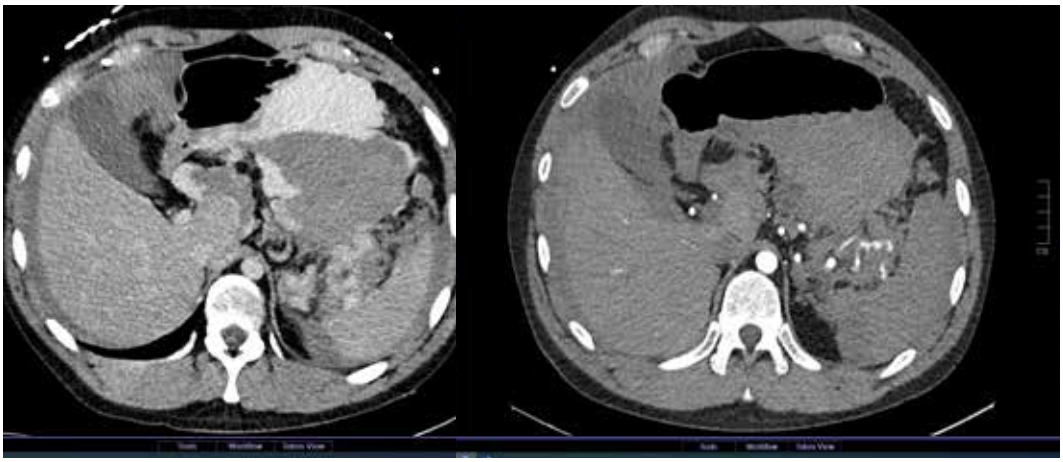

Figure 1 *CT scan at admission.*

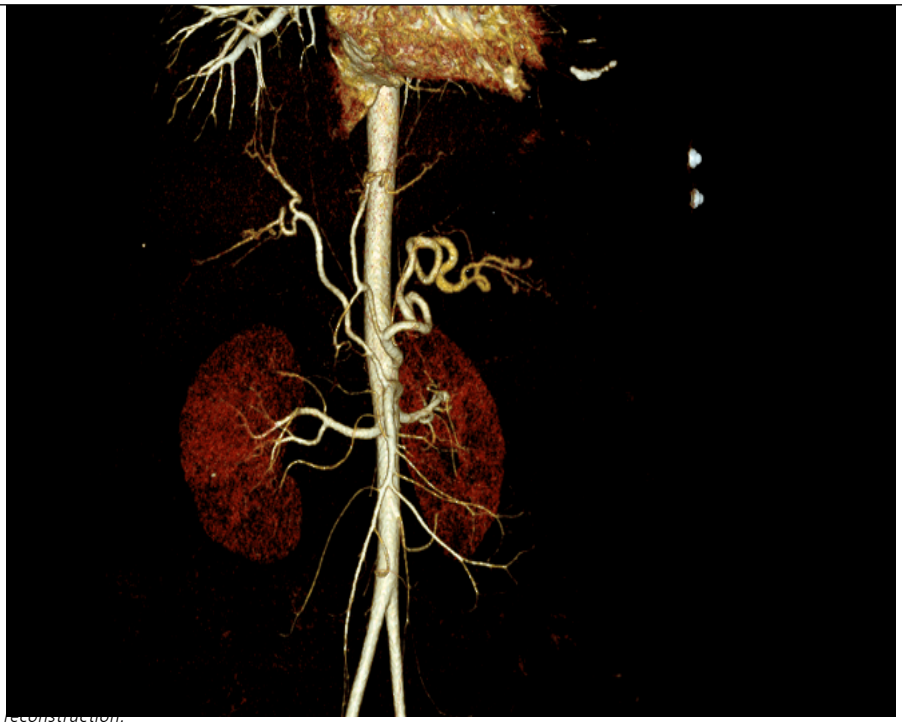

Figure 2 *CT scan reconstruction.*

Bedside ultrasound demonstrated moderate perisplenic and pelvic hemoperitoneum.

Following initial stabilization, contrast-enhanced CT (Figures 1 and 2) revealed moderate hemoperitoneum and active contrast extravasation from the distal splenic artery near the splenic hilum.

A multidisciplinary discussion involving General Surgery, Vascular Surgery, and Intensive Care Medicine recommended an endovascular approach as the safest option, given the patient's underlying condition.

The patient underwent splenic artery embolization via a left humeral artery approach using a 4-French introducer. After selective catheterization of the splenic artery, a 5-French sheath was positioned approximately 2 cm beyond the artery's origin. Arteriography confirmed active bleeding from the distal splenic artery (Figure 3).

Multiple attempts to advance a 5-French support catheter closer to the rupture site were unsuccessful due to marked arterial tortuosity. A stable position was achieved in the proximal third of the artery, and a 2.7-French Terumo Progreat microcatheter was advanced into the medial third. However, it was not possible to reach the rupture site. From this position, distal embolization was performed using 7- and 8-mm coils, followed by embolization of the medial

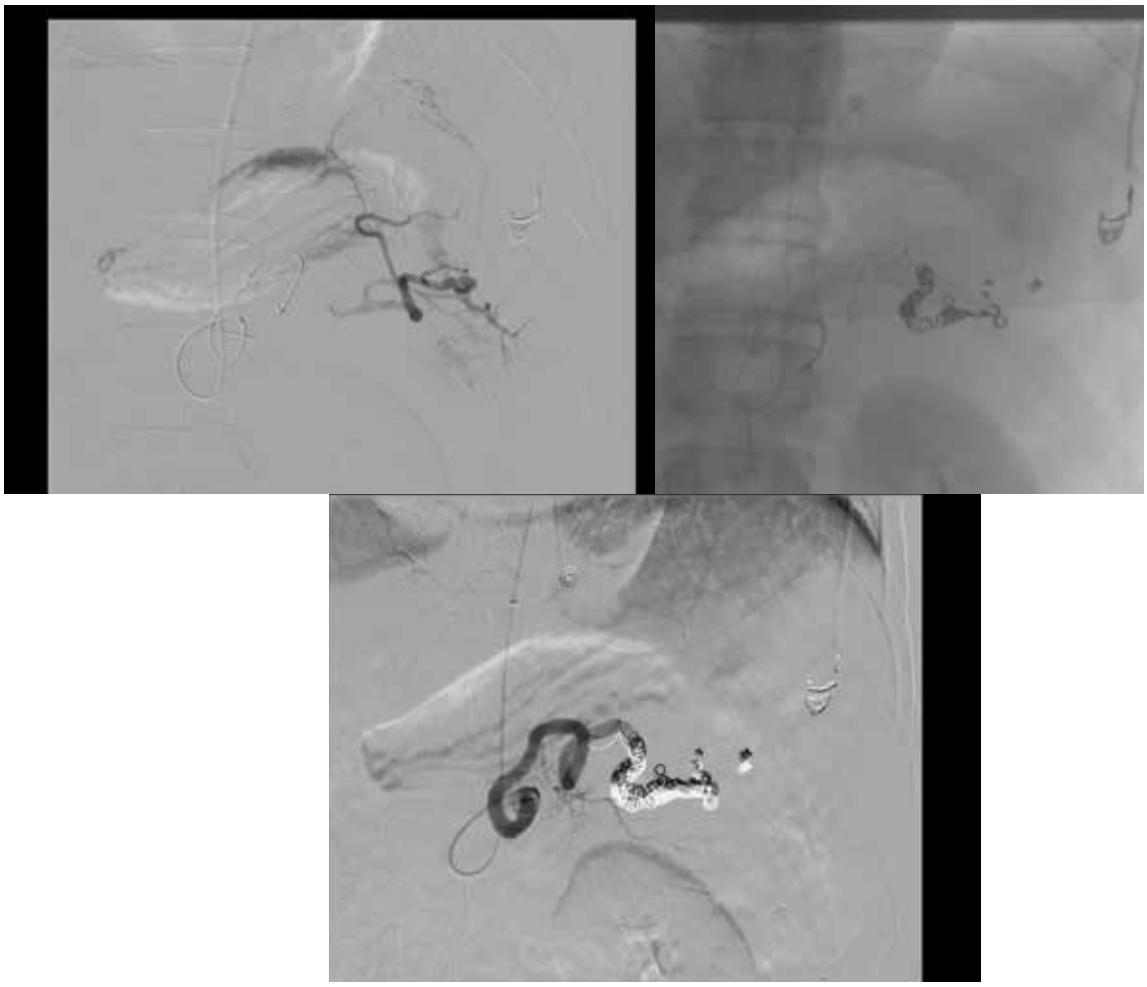

Figure 3 *Coil embolization of the splenic artery.*

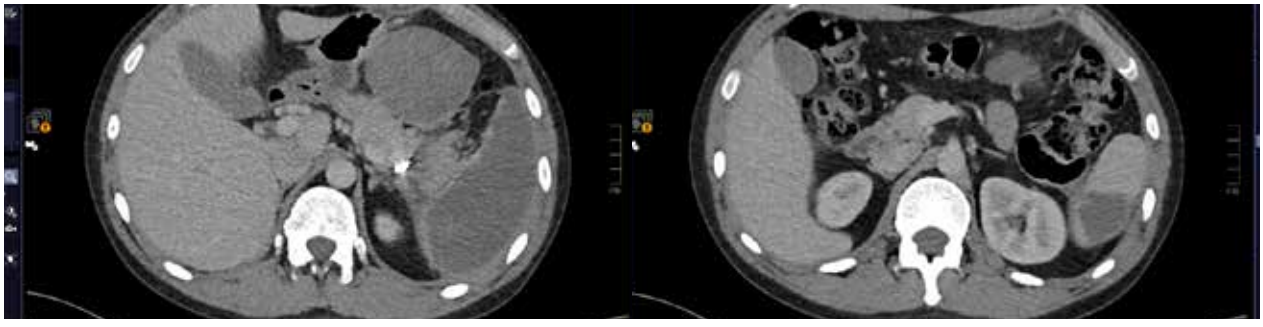

Figure 3 *Control CT scan at discharge.*

splenic artery using 10- and 12-mm coils. Final angiography confirmed complete occlusion of the splenic artery with no further contrast extravasation.

The patient exhibited immediate improvement in hemodynamic stability and transfusion requirements and was admitted to the Intensive Care Unit for continued monitoring. Follow-up CT demonstrated partial splenic infarction without active bleeding (Figure 4).

He was discharged on day 27 after completing post-splenectomy vaccination protocols because of presumed hyposplenism.

DISCUSSION

vEDS is characterized by defective type III collagen production, predisposing affected individuals to spontaneous arterial dissections and ruptures at a young age. When assessing acute abdominal pain in a vEDS patient, vascular complications must always be considered.[6]

Spontaneous splenic rupture is rare but life-threatening, with only isolated cases reported[7-9]. Traditionally, arterial rupture has been managed surgically. However, in vEDS, operative repair carries a substantially increased risk of uncontrollable hemorrhage because of extreme vessel fragility. Endovascular treatment offers the advantage of minimizing vascular manipulation and is often life-saving in this population.[10]

Embolization has increasingly become the preferred strategy for controlling arterial bleeding in vEDS, with observational studies reporting acceptable success rates and fewer complications compared with surgery[9]. Embolization typically involves selective occlusion of the affected artery using coils or other embolic agents, controlling hemorrhage while preserving as much organ function as possible.[3,4]

Technical considerations are crucial in vEDS: minimizing vascular access punctures, using the smallest feasible sheaths, and advancing guidewires and catheters gently to avoid iatrogenic dissections or perforations.[4,10]

Splenic artery embolization may result in infarction, abscess formation, pancreatitis, paradoxical embolization, or vascular access complications[7]. A large retrospective series of 88 vEDS patients undergoing endovascular procedures demonstrated that, despite a relatively high risk of splenic infarction or re-bleeding, mortality was significantly lower than with surgical repair.[3]

Another important aspect is the high recurrence rate of arterial events. Even after successful treatment, vEDS patients remain at elevated risk for new dissections or ruptures. Evidence suggests that recurrence commonly occurs within a few years of the initial event[9].

Follow-Up Considerations

Despite the recognized need for long-term surveillance in vEDS, there is no consensus in the literature regarding the optimal imaging interval after an arterial rupture or embolization. Practices vary widely between centers, and recommendations remain based on expert opinion rather than evidence.

Pharmacological Prevention

Both the ESVS and ESC guidelines recommend celiprolol for vEDS patients, as observational data suggest a reduction in arterial events. Although evidence remains limited and no randomized trials exist beyond the BBEST study, celiprolol is widely considered a reasonable preventive therapy in this population. Its role should be discussed with each patient as part of long-term management.[11-12]

CONCLUSION

Managing spontaneous arterial rupture in vEDS remains a major clinical challenge. Endovascular embolization provides a safe and effective alternative to open surgery, particularly when extreme tissue fragility renders surgical intervention hazardous. Nevertheless, this approach is not without complications, and long-term vigilance is required because of the high recurrence rate of arterial events in vEDS. The absence of evidence-based surveillance intervals further emphasizes the need for individualized follow-up. A multidisciplinary approach is essential to optimize outcomes for these highly complex patients.

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Conflict of Interest

The authors declare no conflicts of interest.

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